The nature of essential tremor

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Summary: The literature concerning essential tremor is confusing. Some accounts describe tremor of a resting type and its accompaniment by other neurological abnormalities. Critical analysis of the pertinent literature leaves some question as to the validity of these observations.

Clinical analysis of 34 patients with essential tremor, personally observed during a four-year period, reveals this to be a monosymptomatic condition. The tremor was usually of both a postural and action type and resting tremor was not observed. There were no other neurological abnormalities. Serious disability was infrequent and, when present, was related to upper limb tremor. Essential tremor should be readily distinguishable from other central nervous system diseases but it may be confused with Parkinson's disease or cerebellar ataxia.

Résumé: *La nature du tremblement essentiel*

La lecture de la littérature sur le tremblement essentiel est troublante. On y trouve certaines descriptions du tremblement essentiel qui se produit au repos et qui est accompagné d'autres anomalies neurologiques. Une analyse critique de la littérature sur ce problème laisse encore en suspens certaines questions sur la validité de ces observations.

Après avoir observé personnellement 34 malades souffrant de tremblement essentiel pendant une période de quatre ans, nous croyons pouvoir affirmer qu'il s'agit d'une pathologie monosymptomatique. Le tremblement était généralement du type postural et actif; le tremblement passif n'a jamais été observé. Nous n'avons jamais noté d'autres anomalies neurologiques. Une invalidité grave, toujours rare, atteignait le cas échéant le membre supérieur. Le tremblement essentiel devrait pouvoir être facilement distingué d'autres pathologies du système nerveux central, mais on risque de le confondre avec la maladie de Parkinson ou l'ataxie cérébelleuse.

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Essential familial tremor, a common nervous system disorder with characteristic physical findings, is often confused with more serious disease of the central nervous system, particularly Parkinson's disease or cerebellar degeneration. The confusion seems to stem from two areas of disagreement. The first concerns the clinical characteristics of the tremor itself and the second relates to the occurrence of neurological abnormalities additional to tremor. In an attempt to clarify these problems a literature review was undertaken and clinical analysis of 34 patients with essential tremor was made.

The nature of essential tremor — resting, postural or action?

In his essay on the shaking palsy James Parkinson[1] made clear distinction between the tremor of Parkinson's disease and other forms of tremor. "In the real Shaking Palsy . . . , the agitation continues in full force whilst the limb is at rest and unemployed; and even is sometimes diminished by calling the muscles into employment". Subsequently, a number of authors have described essential tremor as occurring at rest and simulating the tremor of Parkinson's disease. A recent textbook of neurology[2] contains the following statements: "The tremor most closely resembles Parkinsons' tremor . . . tremor is present at rest and may be inhibited very transiently by volitional activity". Cooper,[3] while recognizing that patients with heredofamilial tremor may present only with intention or postural tremor, states that "still others may present a constant or resting tremor indistinguishable from that seen in parkinsonism". In an extensive review of the subject M. Critchley[4] says "the tremor may appear while the limb is at rest (static tremor or rest tremor) and may be temporarily inhibited during the execution of a voluntary act". However, he immediately goes on to say that "in other cases the tremor seems to be present at rest so long as the hands are outstretched, but ceases when the hands are supported adequately on a table", thus illustrating some ambiguity in the use of the term "resting". Larsson and Sjögren[5] in a survey of essential tremor in a large clinical and genetic population describe it simply as "usually a rest tremor, regular, with a frequency ranging from 5 to 10 per second". In the few clinical descriptions given in their paper, the tremor is often described as increasing markedly with emotion and effort.

The majority of authors consider essential tremor to be of a postural variety and absent at rest. This point of view is summarized by Marshall[6] who believes that essential tremor may be exaggerated during precise movements but that this is not a notable feature.

Perhaps a major part of the disagreement about tremor is semantic. Some writers do not define rest tremor while others clearly treat the terms rest and postural as synonymous.

Is essential tremor a monosymptomatic condition?

Most observers describe essential tremor as being unassociated with other neurological abnormalities but there have been some notable exceptions. In a recent paper E. Critchley[7] records involvement of the lower extremities in 15 of 42 patients and gives a number of illustrative case histories. The following are clearly examples of patients with various types of central nervous system disease of which tremor happened to be one symptom. They cannot therefore be included in the syndrome of essential tremor. The first example is a 67-year-old man with a "masklike expression" in whom "at rest all 4 limbs were on the move". One supposes this represents the development of parkinsonism in a patient with essential tremor, an infrequent sequence which has been recorded before. With two common nervous-system disorders such an association may be expected occasionally. He goes on to describe a 45-year-old man with a gait disorder of 20 years' duration. This patient complained of inversion of the right foot when walking and he had had to reinforce his footwear with steel bars. Examination showed a mild rapid tremor of the right arm and increased tone in the right arm with synkinesis. This appears to be a patient with dystonia. One further case history is worthy of comment. This was a 56-year-old clerk with essential tremor and ataxia of gait. This patient had a previous history of transient double vision and an attack of paraplegia from which he recovered completely. One would agree with the author "that it is probably advisable to regard him as having a double diagnosis of essential tremor and multiple sclerosis".

Two of the patients in Critchley's series had clubbed feet and one had

evidence of peroneal muscular atrophy, suggesting to the author a continuity with other central nervous system degenerations. This suggestion has been raised by others. M. Critchley and Greenfield[8] felt that in some cases essential heredofamilial tremor might represent a *forme fruste* of one of the presenile cerebellar atrophies and discussed this in particular relationship to olivopontocerebellar atrophy.

The paper of Larsson and Sjögren[5] is often quoted in support of the view that essential tremor is more than a monosymptomatic disease. On the basis of observations in 17 of 81 clinically examined cases and historical data in five other cases these authors came to a firm conclusion that essential tremor "cannot generally be labelled monosymptomatic". Their personally examined cases can be classified into three groups. Group I consists of six patients with abnormal signs but no symptoms. The sign is similar in every case and is always described as "varying rigidity" in one or several limbs. Group II includes eight patients with symptoms and signs or symptoms alone. There is a remarkable uniformity in this group as well. The symptom is always "stiff gait" and the signs, as above, varying rigidity of the limbs and occasionally pill-rolling tremor. Rigidity in the upper limbs is never a symptom and "stiff gait" appears to bear little relationship to the distribution of muscle rigidity; for example one patient had stiff gait and slight varying rigidity in the arms, another had stiff gait and tremor of the hands, sometimes of a pill-rolling type, but no rigidity. Group III includes three patients aged 88, 88 and 90 years. Two were suffering from senile dementia and the third, a 90-year-old woman with difficulty walking and stiff gait, had "considerable arthrosis deformans" of the knees. These appear to be patients in whom a double diagnosis would be appropriate.

The evidence is unconvincing that essential tremor is more than a monosymptomatic disease. In a number of reported cases there is merely the association of essential tremor with another disease. In a further number of cases attempts have been made to link essential tremor with conditions such as cerebellar degeneration or torsion dystonia, the suggestion being that essential tremor may represent a *forme fruste* of these disorders. Documentation of such an association is lacking. It is just as logical to consider that tremor may be the initial manifestation of a number of central nervous system diseases, both of a sporadic and familial nature, as to believe that essential tremor may become these diseases by a prolonged process of evolution.

Table I—Clinical data

Case no.	Sex	Age of onset	Duration of symptoms (years)	Tremor: Postural	Tremor: Action	Tremor: Rest	Site of involvement	Tremor ↓ by alcohol	Family history
1	M	50	9	+	+	−	Upper limbs	+	−
2	F	35	10	+	+	−	Upper limbs, mainly right; head	−	−
3	F	29	25	+	+	−	Upper limbs, action right side only; head	−	−
4	M	5	58	+	+	−	Upper limbs	−	+ daughter
5	M	58	5	+	+	−	Upper limbs	−	−
6	M	16	2	+	+	−	Upper limbs	−	+ father
7	M	27	20	+	+	−	Upper limbs; head	+	−
8	F	15	18	+	+	−	Upper limbs; head	+	+ mother and twin sister
9	M	1	21	+	+	−	Upper limbs; head	−	+ maternal grandfather
10	F	32	9	+	+	−	Right hand only	+	+ mother
11	M	44	5	+	+	−	Upper limbs; head	+	−
12	M	56	2	+	+	−	Upper limbs	−	+ two brothers
13	M	47	3	+	+	−	Upper limbs	−	−
14	F	69	1	+	+	−	Upper limbs; head and tongue	−	+ father and daughter
15	M	49	10	+	+	−	Upper limbs; head	+	+ brother
16	M	12	4	+	+	−	Upper limbs; head		+ two maternal uncles
17	F	71	5	+	+	−	Upper limbs; head	−	−
18	M	25	45	+	+	−	Upper limbs; lower limbs (minimal)	−	−
19	F	26	50	+	+	−	Upper limbs	−	−
20	F	14	40	+	+	−	Upper limbs; head	−	−
21	M	15	21	+	+	−	Upper limbs	+	−
22	M	34	25	+	+	−	Upper limbs (right-sided only for many years)	−	+ father
23	M	35	5	+	+	−	Upper limbs; head	+	−
24	M	20	54	+	+	−	Upper limbs; head	−	−
25	F	1	50	+	+	−	Upper limbs, mainly left; head	+	−
26	M	26	30	+	+	−	Upper limbs; head; perioral	+	−
27	F	20	31	+	+	−	Upper limbs; head	+	+ twin brother, older brother
28	M	31	20	+	+	slight	Upper limbs; head	−	−
29	M	65	10	+	+	slight	Upper limbs	−	+ father, sister, brother
30	M	20	43	−	+	−	Upper limbs	−	−
31	F	62	10	−	+	−	Upper limbs	−	−
32	F	52	3	−	+	−	Upper limb, right only	−	−
33	F	15	3	−	+	−	Upper limbs (history only)	−	+ paternal grandfather and father
34	F	69	5	+	−	−	Upper limbs; head	−	−

Table II—Age of onset of symptoms (by decade)

Decade	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79
No. of patients	3	6	8	5	3	4	4	1

The patients described by Larsson and Sjögren represent yet another group. Some of their patients had signs in search of symptoms and most of the others had symptoms without an appropriate physical sign! Almost all of their patients with gait disturbance were more than 70 years old. The authors take cognizance of this fact in their discussion if not in their conclusions: "Rigidity and gait disturbance of the type observed in a considerable number of the tremor cases may occur in advanced age without connection with essential tremor".[11] In a much earlier paper M. Critchley[9] stressed this same point: "But most of these numerous little signs and manifestations, the characteristic gestures, attitude, stance, and gait of the elderly which so obviously distinguish them from the youthful, are really manifestations of an extrapyramidal order". It may well be considered, in the aged population with essential tremor, that associated extrapyramidal signs are part of the normal senium. It is only in this group with "senile tremor" that parkinsonism is likely to enter truly the differential diagnosis of essential tremor.

Clinical material

Thirty-four patients (20 men and 14 women) were examined during a four-year period. The pertinent clinical data are recorded in Table I.

History

The age of onset of essential tremor is tabulated by decade (Table II), the results being similar to those found in other series.

The duration of symptoms at the time of initial examination was always considerable. This emphasizes the benign nature and slow progression of the disease. In 20 of 34 patients tremor had been present for more than 10 years. The usual history was of long periods during which tremor worsened either very slowly or not at all. In a few patients fairly rapid progression of symptoms took place after an extended period of non-progression.

In most patients disability was mild and insufficient to warrant treatment. A number were referred with the incorrect diagnosis of Parkinson's disease and some had been on antiparkinsonian medication. Reassurance was often all that was necessary in these cases. Many patients complained of embarrassment due to head or hand tremor and most complained of deterioration of handwriting. Almost without exception, worsening of tremor during and following activity or while under observation was reported. It is an old observation and one characteristic of this condition, that alcohol, even in small amounts, may produce considerable reduction in tremor. This history was obtained in 11 patients.

The tremor begins either in the head or hands. The involvement of the upper limbs is often asymmetrical and occasionally unilateral.

Significant functional disability was seen in only four patients. In each case it was due to upper limb involvement by postural and action tremor, so severe as to interfere with occupation or activities of daily living, such as eating.

A positive family history was obtained in 13 patients. The pattern of involvement (see Table I) indicates an autosomal dominant mode of transmission.

Physical findings

In no case was a neurological abnormality other than tremor detectable nor was there any instance of involvement of the lower limbs. In cases 1 to 29 a combination of postural and action tremor occurred. Postural tremor is seen when the hands are held outstretched, appearing almost at once and worsening in many cases as the position is maintained. The tremor is generally coarse and may be compound. Sometimes "pill-rolling" elements were seen. Persistence or worsening of tremor during the performance of the finger-to-nose-to-finger test was noted in all these patients. The tremor is apparent throughout movement and worsens as the target is approached. It most closely resembles the tremor of "cerebellar" disease but differs from it in that hypermetria does not occur. Rest tremor was notably absent, being recorded to a slight degree in two patients only (cases 28 and 29).

The head tremor is also of a postural variety and disappears when the head is supported. It may occur in either the horizontal or vertical plane. It is intermittent in the earlier stages and, like the limb tremor, is worsened by activity and emotional upset. Head tremor may precede limb tremor by several years or may follow limb tremor by equally long periods of time. A combination of head and arm tremor is common and was seen in 17 of 23 patients in this series.

Cases 30 to 34 differed in having only action tremor (four patients) or only postural tremor (one patient).

Discussion

Essential familial tremor, as seen in this series, is a monosymptomatic condition. The tremor is of a postural variety, persisting or worsening during action. Rarely a purely postural or a purely action tremor is seen. Rest tremor as an isolated phenomenon was never encountered and probably does not occur if the limb is fully supported and completely relaxed. As a consequence, the differentiation between Parkinson's disease and essential tremor should pose no great diagnostic problem. Essential tremor bears much more resemblance to cerebellar tremor but differs from it in two important respects: in essential tremor there is no dysmetria and ataxia is not seen in the lower limbs, even when the arms are so involved as to interfere with activities of daily living.

Incorrect diagnosis of this common condition is unfortunate for it leads not only to inappropriate treatment but to unnecessary mental distress in the patient who fears he may have a progressive and crippling disease. In many instances essential tremor needs no treatment. Recent reports[10,11] on the use of the beta-adrenergic blocking agent propranolol, are encouraging and in patients with severe disability excellent results have been achieved with stereotaxic surgical procedures similar to those used in the treatment of Parkinson's disease.[12,13]

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